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Case report

Cystic fibrosis and COVID-19: Care considerations

Alicia A. Mirza [a], Elika J. Rad [b,*], Paul K. Mohabir [a]

[a] Stanford University School of Medicine, Stanford, CA, USA
[b] Stanford Health Care, Stanford, CA, USA

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ABSTRACT

The coronavirus disease 2019 (COVID-19) pandemic has demanded large scale changes in patient care. People with cystic fibrosis have unique considerations, including underlying lung disease and routine aerosolizing therapies, but there is insufficient evidence to create comprehensive practice guidelines. We share a case of a patient with CF and COVID-19 as well as alterations to routine CF care at a large academic center. Key considerations include accessible COVID-19 screening, augmented infection control practices, and rapid integration of telemedicine.

1. Introduction

Clinical manifestations of coronavirus disease 2019 (COVID-19) are widely variable from asymptomatic to severe illness with progression to acute respiratory distress and respiratory failure [1]. Age, comorbidities, and male sex have been associated with worse outcomes in the general population with COVID-19, but less is known about its effects specific to cystic fibrosis (CF) [2]. Historically, patients with CF have had an increased risk of complications from respiratory viral infections such as the 2009 H1N1 influenza virus, raising the question if this elevated risk would also occur with COVID-19 [3]. Additionally, routine CF care including pulmonary exacerbation management and airway clearance therapy is complicated due to the transmissibility of the virus. We present a case of COVID-19 in a patient with CF who returned to baseline function and outline important steps in routine CF care.

2. Case report

The patient is a 29-year-old man with a history of CF with moderate obstructive lung disease, exocrine pancreatic insufficiency and gastroparesis. His genotype is 394delTT/G1061R with (TG)10-7T/(TG)10-9T poly T tract repeat and is not eligible for CFTR modulator therapy. Respiratory cultures chronically grow multidrug resistant pseudomonas and methicillin-resistant staphylococcus aureus (MRSA). He presented with chest pain, shortness of breath, but did not have increased sputum as was typical of his prior CF exacerbations. Chest radiograph showed upper lobe predominant bronchial wall thickening and bronchiectasis with nodular and interstitial opacities in the left greater than right bilateral lungs. COVID-19 nasopharyngeal reverse transcriptase-polymerase chain reaction (RT-PCR) testing was positive. As this was early in the pandemic, he was enrolled in a clinical trial and randomized into either receiving remdesivir or placebo. He was treated with a 14-day course of meropenem and aztreonam based on prior respiratory culture sensitivities. The patient was discharged home after 6 days and received the majority of his IV antibiotics at home. He continued to have chest pain and shortness of breath, and hospital cultures showed a new meropenem-resistant pseudomonal species, thus he was broadened to ceftolozone-tazobactam with inhaled colistin for two additional weeks. Symptoms improved approximately two weeks after finishing antibiotic therapy.

3. Discussion

Available data about COVID-19 in patients with CF is limited. At the onset of the pandemic, presumptions were made that these patients would have a worse prognosis. This was evidenced by several states creating emergency regional triage plans that included the presence of CF with decreased lung function (FEV1<30%) as a reason for denying intensive care unit admission, (which was later contested by CF experts) [4]. The most comprehensive case series to date describes the presentation of 40 patients with CF and confirmed COVID-19 collected from CF registry data from eight countries [5]. There were no specific risk factors for the contraction or severity of illness detected in this heterogenous group. Their disease manifestations and duration of illness did not seem to differ greatly from the general population. Only one of 40 patients required invasive ventilation, and this patient was post-lung

* Corresponding author. Adult Cystic FibrosisCenter for Advanced Lung Disease, Stanford Health Care,300 Pasteur Drive, M/C 5351, Stanford, CA 94305, USA.
E-mail addresses: Alicia.a.mirza@gmail.com (A.A. Mirza), ERad@stanfordhealthcare.org (E.J. Rad).

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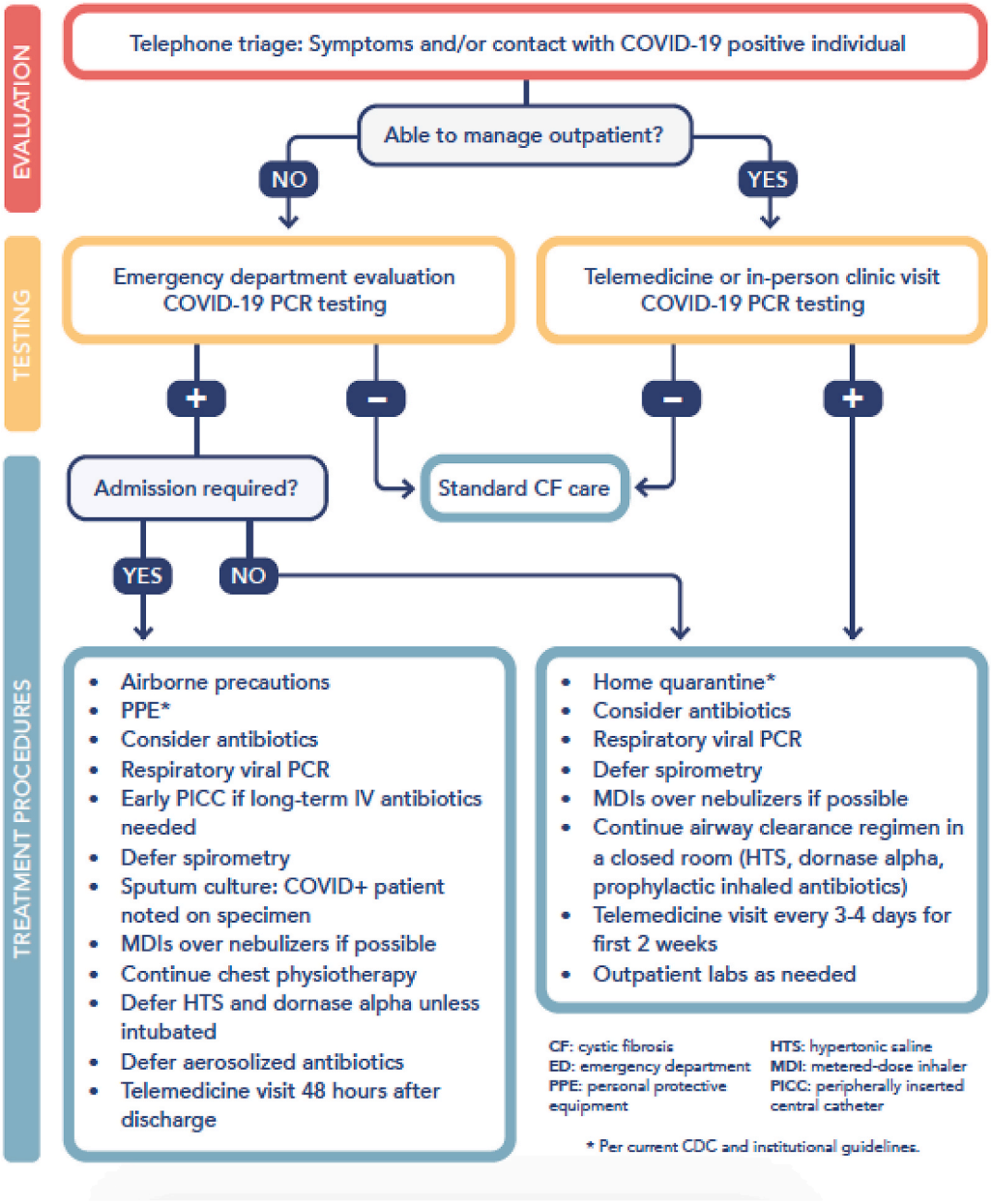

Fig. 1. Proposed Algorithm of Routine CF Care Considerations if COVID-19 Suspected.

transplantation. A retrospective observational study of in Spain found a lower incidence and mortality in 8 CF patients with COVID-19 when compared to the general Spanish population [6]. Therefore, available data in a small number of patients with CF does not show a significantly worse prognosis despite their underlying lung pathology. There is a notable amount of variability amongst the cases in terms of severity of disease, co-infections, genotype and treatments received. Long-term effects of infection are still unknown.

While early data is limited, it is clear that COVID-19 respiratory exacerbations require alternations in routine CF care. There are no comprehensive practice guidelines on management of COIVD-19 in cystic fibrosis at this time. Fig. 1 depicts our proposed algorithm based on clinical experience and available guidelines. It has been argued that the dry cough of COVID-19, along with fever and myalgias, will distinguish it from typical CF exacerbation symptoms and from other pathogens [3]. This was true in our patient, however many patients on CFTR protein modulators do not have increased sputum with routine exacerbations thus making testing essential for diagnosis. Upon symptom reporting, CF nurse coordinators perform telephone screening and RT-PCR testing if indicated.

Special consideration does need to be taken for airway clearance therapies in the hospital setting. In general, viruses spread through direct or indirect contact, respiratory droplets and fine-particle aerosols (droplets and droplet nuclei with aerodynamic diameter $\leq$5 μm) [7]. Cough-inducing procedures such as chest physiotherapy vesting, nebulized hypertonic saline and dornase alfa are important components of CF care, but risk aerosolization of viruses during these treatments. In patients with known or suspected COVID-19, metered dose inhalers (MDIs) are preferred over nebulized therapies whenever possible. While chest physiotherapy needs vary among patients with COVID-19, this therapy should be continued for all patients with CF with appropriate isolation precautions. Threshold for antibiotics may be lower considering endobronchial colonization and potential limitations in airway clearance therapy. Home IV antibiotic therapy can decrease length of stay. Regular follow up with telemedicine visits allow for more frequent home monitoring.

4. Conclusion

Early data shows patients with cystic fibrosis have similar disease manifestations and disease severity to the general population despite underlying lung disease, as was exemplified by our case. The outlined treatment algorithm can aid in diagnosis and treatment of these patients. There are many other important considerations, such as public health measures and treatment options for COVID-19 that are active areas of ongoing research. CF Centers should be sharing their clinical experiences in this pandemic to optimize patient care and safety.

Declaration of competing interest

No conflicts of interest to disclose.

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Figure by Covid Creatives.

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